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Misleading Leads: Wilms Tumor With a Metachronous Lesion Appearing in the Remaining Kidney

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Medullary dysplasia may be present in the kidneys of children with the Beckwith-Wiedemann Syndrome (BWS). This is usually visible only at the microscopic level, but superimposed pyelonephritis in a child with the BWS led to gross changes that produced a filling defect on CT scanning of the remaining kidney. The finding could have been misinterpreted as a metachronous wilms tumor. Med. Pediatr. Oncol. 30:180–182, 1998.

Key words: Wilms tumor; Beckwith-Wiedemann syndrome; pyelonephritis; renal cortical atrophy

INTRODUCTION

A 6-month-old girl was found to have Beckwith-Wiedemann syndrome (BWS) after hemihypertrophy was identified at birth [1]. An abdominal ultrasound performed at that time showed a mass in the right renal hilum which on CT scanning was 6.8 × 4.2 × 5.2 cm in size. The left kidney was normal, and the chest showed no evidence of disease. An abdominal ultrasound examination had been performed when the child was 2 days of age because of a mass palpable in the left abdomen, but this revealed normal kidneys on both sides.

A wedge biopsy was performed to facilitate a renal-sparing approach. This was because the overgrowth syndromes (hemihypertrophy and BWS) are associated with nephrogenic rests; hence, these patients are at increased risk of developing bilateral Wilms tumors [2,3]. The bi-

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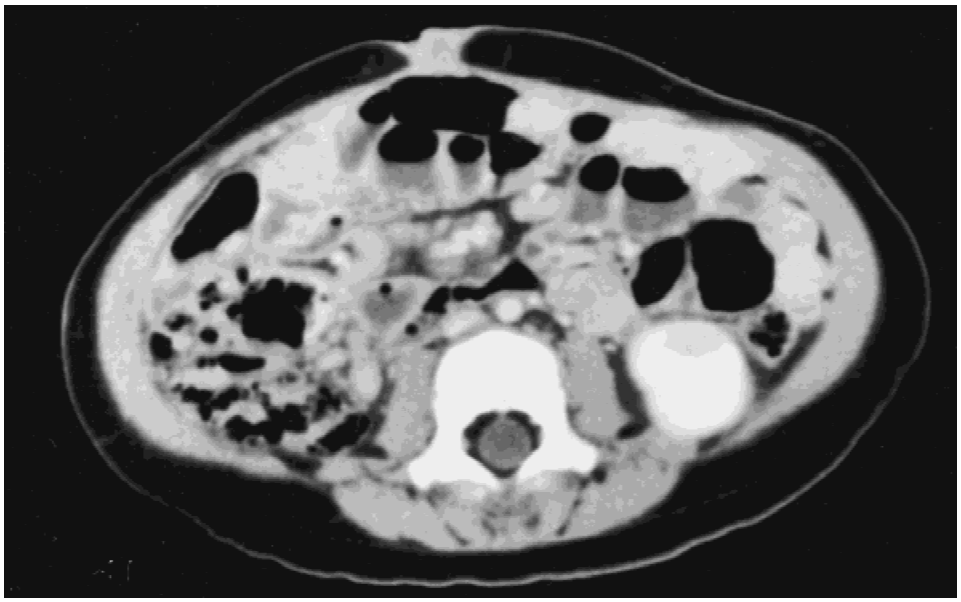

Fig. 1. There is an oval defect anteriorly in the remaining opacified left kidney. It compresses the underlying major calyx. No other filling defects or exophytic lesions are visible.

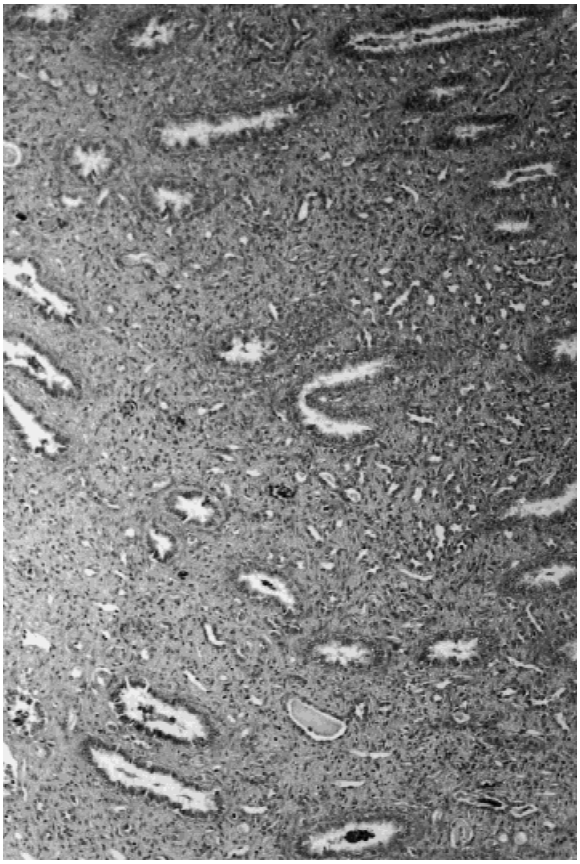

Fig. 2. Medullary dysplasia. Collecting ducts are widely separated by a mesenchymal background. This appearance is caused in part by incomplete penetration of Henle's loops into the deep medulla, and is a common feature of kidneys from children with BWS.

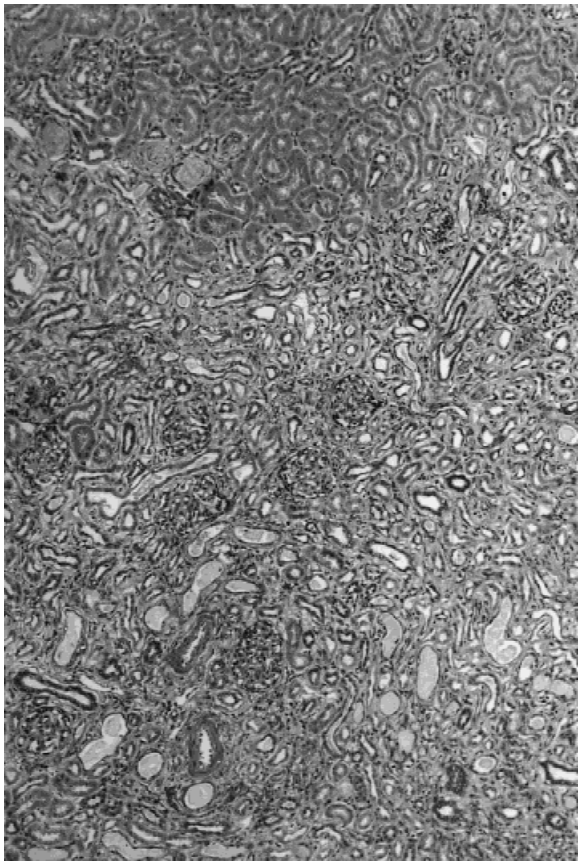

Fig. 3. Renal cortex overlying dysplastic medulla shown in Figure 2. At the top edge of the field, cortical tubules with tall epithelium are seen. The tubules in the remainder of the field are atrophic, and many tubule lumens contain casts. It is not clear whether this appearance reflects local obstruction, or the effects of recent cortical ischemia from an unknown cause.

opsy revealed favorable histology Wilms tumor. The left kidney was normal to inspection, and all nodes removed were free of disease. The patient was then treated with vincristine and dactinomycin. She showed a good response, the tumor size 3 months later being 3.3 × 2.5 × 2.3 cm. A partial nephrectomy was then performed, but because of positive margins, a right total nephrectomy was necessary. Pathology again showed favorable histology Wilms tumor without nephroblastomatosis or other nephrogenic rests being identified. Chemotherapy was continued for 4 months, and 6 months after diagnosis a CT scan showed no evidence of disease in the tumor bed or in the left kidney.

A routine follow-up CT scan was performed 3 months later when 5 months off-therapy showed a 1.0 × 0.8 × 0.9 cm lesion in the lower pole of the left kidney. Since the kidney had appeared normal prior to this examination, a metachronous Wilms tumor or an enlarging nephrogenic rest were prominent in the differential diagnosis (Fig. 1). Several expert opinions were solicited at that time, which included recommendations for the initiation of chemotherapy, potentially including doxorubicin, close follow-up observation with scans, or biopsy.* Biopsy proof was considered necessary by the responsible physicians, and laparotomy confirmed the presence of a palpable lesion on the left side. A wedge biopsy was done, and this showed localized tubular atrophy and the medullary dysplasia seen in BWS, with a superimposed chronic pyelonephritis (Figs. 2 and 3).

Beckwith described the dysplasia of the renal medulla seen in Figure 2 in his original description of the Beckwith-Wiedemann syndrome [1]. These changes are ordinarily found at the microscopic level. They became identified in this case because of the confounding additional pathology that produced the gross defect visible on CT scanning. No further therapy was given, and the patient remains free of disease 1 year

*Editor's note: I was consulted at this point, and recommended the initiation of chemotherapy, asking the question, ''What else could this be?'' Dr. Hilden and colleagues and Dr. Beckwith have shown us what else it could be.

from the time of diagnosis. She is being followed by scans.

CONCLUSION

Changes unrelated to neoplasia will sometimes be detected by regular imaging of the kidney in children at risk for Wilms tumor. The sensitivity of modern technologies has led to detection of nephrogenic rests, focal infection or ischemia, cysts, localized renal dysplasia, and other nonmalignant conditions (Beckwith, personal communication). Repeated imaging to determine the growth rate of a suspicious lesion may sometimes be preferable to immediate surgical intervention.

REFERENCES

1. Beckwith JB: Macroglossia, omphalocele, adrenal cytomegaly, gigantism, and hyperplastic visceromegaly. Birth Defects OAS 5:188–196, 1969.
2. Blute ML, Kelalis PP, Offord KP, et al.: Bilateral Wilms tumor. J Urol 138:968–973, 1987.
3. Beckwith JB: Precursor lesions of Wilms tumor: Clinical and biological implications. Med Pediatr Oncol 21:158–168, 1993.